Giant pedunculated esophageal liposarcoma associated with Hurthle cell thyroid neoplasia

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ABSTRACT

Giant pedunculated liposarcoma of the esophagus is considered a rare entity. They impose difficulties in management, especially the surgical approach. We report a case of giant pedunculated liposarcoma of the esophagus in a 62-year-old male who underwent cervical excision of this large tumor, and made a full recovery. Hurthle cell thyroid cancer was found during the work-up, which was managed accordingly.

Case Report. This is a case of a 62-year-old male who was referred with complaints of progressive dysphagia and choking sensation over the last 6 months. Significant weight loss, 10 Kg in 3 months, have been reported. No history of neurological or musculoskeletal symptoms. His past history did not reveal any previous surgeries or comorbidities. The rest of his medical history and systemic review was not contributory. His physical exam revealed average built male, normal vital signs, and no evidence of clubbing or lower limb edema. A battery of laboratory investigations were requested, and complete blood count, renal profile, hepatic profile, bone profile and serologies were within normal limits. A posteroanterior and lateral chest x-ray (CXR) revealed a retrocardiac shadow. Chest CT scans revealed hypopharyngeal and upper esophagus soft tissue mass extending to the gastroesophageal (GE) junction. There was no esophageal wall thickening or mediastinal lymphadenopathy, however the esophagus was markedly dilated (Figure 1a and Figure 1b). A right thyroid nodule was seen and it was worrisome. The positron emission tomography (PET) scan showed a large mixed density esophageal mass with only activity in the most inferior part (Figure 2a) and activity in the thyroid gland (Figure 2b). An upper GI endoscopy (UGE) revealed a small soft tissue mass component in the pharynx and the large component occupying the esophageal lumen.
whole length of the esophagus until the GE junction. It was a large pedunculated polyp separated from the esophageal wall. The decision was to excise the polyp surgically. The cervical approach was elected since it addresses both pharyngeal, as well as the esophageal components of the mass. Under general anesthesia, single tube ventilation and supine position, the UGE was repeated to exactly localize the small peri-epiglottic component. Neck and anterior chest scrubbing was carried out. The neck was explored through a left oblique neck incision parallel to sternomastoid muscle. Dissection reaching the esophagus proceeded with adequate dissection around it, preserving the recurrent nerve. The esophageal muscle and mucosa were opened at the left anterolateral cervical part. The polyp was identified, as well as the stalk, and it was delivered completely through the cervical incision with difficulty. The stalk base was stapled by Endo GIA. The large component was 24 cm in length (Figure 3). The smaller component was dealt in the same fashion. The esophagus was repaired after advancing a size 50 Bougie with 3/0 PDS sutures. Nasogastric tube (NGT) was advanced after removing the Bougie. The closure was carried out in the usual fashion. We elected to insert a drain, which was removed in the second day post-operatively. The patient had an uneventful post-operative course, and a contrast swallow study revealed no leak. He resumed swallowing and his dysphagia resolved. The FNAC for the right thyroid lesion came consistent with Hurthle cell carcinoma, which was managed accordingly by the endocrine surgeon after complete recovery. The final esophageal mass histopathology came consistent with atypical lipomatous well-differentiated liposarcoma. The patient was followed after 6 months and one year with no evidence of recurrence.

Discussion. The first reported case of esophageal liposarcoma in the English literature was in 1983 by Mansour et al. Combined esophageal sarcomas accounts for 0.5% including leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma, and liposarcoma. Gastrointestinal liposarcomas are rare accounting for 0.1-5.8% of all esophageal tumors, and it is mainly in distal ileum and large bowel. Even rarer, is the esophageal liposarcoma accounting for 1.2-1.5%.
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It has a male predominance of male to female ratio, approximately 1.16:1, with age range of 43-73 years (mean; 58.7 years). It is categorized into the following histological types: 1. well-differentiated; 2. myxoid; 3. pleomorphic; 4. dedifferentiated; and 5. round cell.3 The most common type is the well-differentiated (70%) followed by the myxoid type (25%), and the remaining accounting for 5%. It has been reported that these subtypes can be found in one case. The polypoid type is the most common (98%), whereas only one transmural case has been reported. It originates from the mucosa or submucosa in the cervical region in 84.6% of cases.1,3 The tumor size range from 4-23 cm (mean; 12.8 cm), and in our case was 24 cm. With regard to tumor genetics, Mutated Double Minute (MDM2) and CDK4 are reported to be positive in liposarcoma. Some reports demonstrated positive S100 immunohistochemistry.2

The MDM2 is a negative regulator of P53. After binding to P53, it enters a pathway with induction of MDM2 leading to shifting the balance towards cell survival instead of apoptosis.7,8 These patients usually present with progressive dysphagia, nausea, throat discomfort, foreign body sensation, vomiting, cough, or weight loss. Some have reported odynophagia, anemia, or even sudden death.9 Recurrent vomiting followed by extrusion of the mass was also reported.10 These polypoid tumors develop small proximally and grow distally in line with peristalsis.5 The radiological findings are helpful but not pathognomonic. The hallmark is low density (fat tissue) shown on CT scans and MRI.1 Barium swallow, esophageal manometry, and endoscopy are helpful in establishing the diagnosis.

Esophageal ultrasonography is helpful in assessing the complexity and nature of the mass.9 It could be confused with other endoluminal lesions, such as megaesophagus, achalasia, or bezoars.10 On MRI, it can be confused with other endoluminal lesions, such as megaesophagus, achalasia, or bezoars.10 On MRI, it can also demonstrate a high signal intensity in T1-weighted images.3 The treatment is excision and options are: transcervical; trans-thoracic; trans-gastric; or endoscopic. Since most of these tumors are endoluminal soft polyypodial tumors, simple excision with transection of the stalk is the basis of treatment. Esophagectomy has been reported in some cases.1,2,10 These tumors are considered radiosensitive, and a room for post-operative radiotherapy has been advised to reduce local recurrence.10 Liposarcoma has been associated with other primary malignant tumors in 12% of cases.9 Association with serous ovarian carcinoma and lung cancer has been reported. Moreover, it has been associated with Li-Fraumeni-like syndromes in limited reports.9,11 We add the association with Hurthle cell thyroid cancer. The prognosis depends on the histologic category, grade, location, and sufficient surgical excision.3 These tumors did not show propensity to metastasize.1 Local recurrence is lower in well-differentiated category with a 5-year survival 75-100% in this category.10 The local recurrence incidence increase in the myxoid, and even worse in the pleomorphic category approximately 80% with estimated metastasis around 29-44%.2,9

In conclusion, esophageal liposarcoma is an extremely rare entity. Surgical excision is still the standard management. These tumors are radiosensitive, therefore, radiotherapy is considered. The prognosis depends on many factors, such as the subtype, grade, and extent of resection.

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References